# Takayasu Arteritis in Southern Sweden

Aladdin J. Mohammad and Thomas Mandl

***ABSTRACT.*** ***Objective.*** To study the epidemiology and clinical characteristics of Takayasu arteritis (TA) in southern Sweden.

***Methods.*** The study area is situated in Skåne, the southernmost county in Sweden (total population December 2011: 983,419, 50.5% women). Patients were identified using clinical registries in all the 5 hospitals and private rheumatology clinics within the study area between the years 1997 and 2011. The diagnosis of TA was confirmed by medical records review. Only patients fulfilling the 1990 American College of Rheumatology classification criteria were included.

***Results.*** Thirteen patients (all women) were identified. The median age at diagnosis was 23 years [interquartile range (IQR) 16–38]. Ten patients were diagnosed between 1997 and 2011. The annual incidence rate was estimated to 0.7/million inhabitants (95% CI 0.3–1.2) and 1.5/million among women (95% CI 0.6–2.4). Patients were followed for a median of 9 years (IQR 4–17.5). As of January 1, 2012, all 13 patients were alive and living within the study area. The point prevalence per million inhabitants was 13.2 (95% CI 6.0–20.4), and 26.2 among women (95% CI 11.9–40.4). Subclavian arteries were the most commonly affected vessels. Organ damage was common, affecting all patients. Seven pregnancies resulting in 5 live births and 2 abortions were registered after the diagnosis of TA.

***Conclusion.*** The incidence of TA in Sweden is comparable to recently reported rates from other European studies, while the prevalence is higher than previously reported. The prognosis of TA is good, but the rate of damage is high. (First Release March 15 2015; J Rheumatol 2015;42:853–8; doi:10.3899/jrheum.140843)

*Key Indexing Terms:*
TAKAYASU ARTERITIS EPIDEMIOLOGY POPULATION-BASED STUDY
OUTCOME PREGNANCY ORGAN DAMAGE

Takayasu arteritis (TA) is a large vessel vasculitis of unknown etiology, predominantly affecting the aorta and/or its major branches[1]. TA has characteristic clinical and epidemiological features, with disease onset usually occurring before the age of 40 years, and with female predominance. The main clinical features include claudication of the extremities, decreased brachial artery pulses, blood pressure differences, and bruit over major arteries in the upper limb[2], but TA has no characteristic laboratory or serological features. The diagnosis of TA depends solely on the demonstration of vessel stenosis and dilatation in various invasive and noninvasive imaging studies. The epidemiological characteristic of TA is a worldwide distribution with an annual incidence rate per million reported to be 0.4 in Denmark[3], 0.8 in the United Kingdom[4], 1 in Germany[5], 2.2 in Kuwait[6], and 2.6 in Minnesota, USA[7]. The prevalence data are limited and reported to range between 4.7 per million in the United Kingdom[4], 7.8 in Kuwait[6], and 40 per million in Japan[8]. In Sweden, the prevalence of TA was described in the mid-1970s (6.4 per million)[9] while the incidence is largely unknown.

Organ damage is still an important problem among patients mainly because of a combination of longterm exposure to toxic therapies such as corticosteroids and chronic vessel inflammation. Biological therapies have shown efficacy in TA, including the anti-tumor necrosis factor-α[10], and more recently, the use of interleukin 6 inhibitors[11]. There is therefore a need for an update on the epidemiology and clinical outcomes of TA in the era of these new therapies.

The aims of our study were to (1) study the annual incidence rate and point prevalence of TA in a well-defined population in southern Sweden; (2) describe the clinical presentation, vascular distribution, pregnancy outcome, and surgical interventions during the course of TA; and (3) estimate the irreversible organ damage by the vasculitis damage index.

## MATERIALS AND METHODS

*The study area and population.* The study area consisted of 3 healthcare districts in Skåne, the southernmost county in Sweden. The total population on December 31, 2011, was 983,419 (10% of the adult population in

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*Supported by grants from the Swedish Rheumatism Association (Reumatikerförbundet), the Swedish Medical Association (Läkarsällskapet), and the Faculty of Medicine, Lund University.*

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*Accepted for publication January 16, 2015.*

Sweden)[12], of which 195,766 were born outside Sweden. The population in the study area increased by 125,547 inhabitants (13%) from 1997 to the end of 2011. The total population aged ≤ 40 years was 470,311 in 2004 (231,516 women). Women made up 50.5% of the total population and 49.2% of the age group ≤ 40 years[12]. The study area is about 5334 km$^2$ (1.2% of the total area of Sweden) and is divided into 22 municipalities, the largest being the cities of Malmö, Helsingborg, and Lund. "Region Skåne" is a regional public body with administrative and financial responsibility for the health of the inhabitants, and for providing medical and dental services. The study area is served by 5 hospitals, all run by Region Skåne: Skåne University Hospital in Lund and Malmö, Trelleborg Hospital, Landskrona Hospital, Helsingborg Hospital, and Ängelholm Hospital.

*Case identification*. The clinical registries including the inpatient and out-patient registries were searched for patients assigned with the diagnosis codes of TA during the study time [*International Classification of Diseases*, 9th ed (ICD-9): 446.7 in 1997, and ICD-10: M31.4 for the period 1998–2011]. Additionally, patients assigned the diagnosis codes of aortitis (ICD-9: 447.6 and ICD-10: I 79.1) were also identified. Patients were identified using the clinical registries at the departments of rheumatology, nephrology, and vascular medicine at Skåne University Hospital in Malmö and Lund, and the Departments of Internal Medicine at Trelleborg, Landskrona, Helsingborg, and Ängelholm hospitals. All private rheumatology clinics within the study area were also asked if they had treated patients with TA during the study time.

*Data collection*. A structured review of all case records was carried out to ascertain the diagnosis of TA. Patients with clinical and radiographic diagnosis of TA were included. Only patients fulfilling the 1990 American College of Rheumatology (ACR) criteria for the classification of TA were included in our study[2]. Data on clinical presentations and laboratory findings at diagnosis were also extracted from case records. In addition, a detailed review of all available radiology reports was done to study the distribution of arterial lesions. At the last followup (December 2013), data on treatments and the vasculitis damage index (VDI)[13] were collected for all patients. The VDI is registered as total VDI score (the sum of all items), system score, critical damage, severe damage, and treatment-related damage as defined by Exley, *et al*[14]. System score referred to the number of systems to which patients were assigned with at least 1 item of damage. Severe organ damage was defined as a total VDI score of ≥ 5, critical damage ≥ 1, or system score ≥ 3. The assessment of VDI was carried out on the date of prevalence estimate, January 1, 2012. Disease duration was time in years from diagnosis to last assessment. The diagnosis delay was defined as time elapsed in months from the first possible symptoms of TA to the date of diagnosis.

Data on pregnancies and their outcomes were collected from time before and after diagnosis of TA.

*Statistical analyses*. Data are presented in median and interquartile range (IQR) or number and percentage unless otherwise stated. The differences between groups were compared using the nonparametric Mann-Whitney U test and chi-square test when appropriate. The p value of $< 0.05$ was considered significant. For the incidence estimates, all newly diagnosed patients with TA between 1997 and 2011 who were living within the study area at the time of diagnosis were included. For the prevalence estimates, all patients with a diagnosis of TA who were living within the study area on January 1, 2012, were included. The incidence rate per million inhabitants for each calendar year during the study period was calculated using the total population each year at January 1 as the denominator and number of patients diagnosed in that particular year as the numerator. Similarly, the incidence rate for the population aged ≤ 40 years was calculated for each year during the study period. For the prevalence estimates, the denominator was the total population on December 31, 2011 (983,419 inhabitants). Statistical analyses was performed using the SPSS for Windows, version 20.0 (IBM SPSS Statistics). The study was approved by the local Ethics Committee at the Faculty of Medicine, Lund University (2010/517).

## RESULTS

The first searches in the diagnosis databases resulted in the identification of 29 patients who were assigned with the diagnosis code for TA and were living within the study area. Case records of patients living outside the study area were not reviewed. The case record review resulted in the identification of 13 patients (all women) with TA diagnosis based on clinical and radiological characteristics. All patients fulfilled the ACR criteria for classification of TA[2]. The reasons for excluding the remaining 16 subjects were as follows: did not fulfill the TA classification criteria (n = 9), patients with isolated aortitis with or without giant cell arteritis (n = 2), case record was not available (n = 1), not enough data were available to make a diagnosis of TA (n = 3), and 1 patient was incorrectly assigned an ICD number for TA. None of the additional 8 people who were assigned the diagnosis codes for aortitis were found to have TA. No cases were identified from the private clinics in the area.

Of the 13 patients who were included in our study, 10 were diagnosed between 1997 and 2011 (incident cases), and the remaining 3 patients were diagnosed before 1997, but were still alive and living within the study area on the date of point prevalence estimation.

The median age at diagnosis was 23 years (IQR 16–38, range 9–61) and at date of point prevalence estimate was 39 years (IQR 28.5–48.5, range 20–68). Two patients were aged 51 years and 61 years at diagnosis, both fulfilled the ACR criteria for TA, and none had any signs or symptoms suggestive of giant cell arteritis. Further clinical and laboratory characteristics are shown in Table 1. Eight patients (61.5%) were of Swedish ancestry, 1 Asian, 2 Arab, 1 African, and 1 patient from northern Europe. All the non-Swedish patients were first-generation immigrants.

*Annual incidence rate*. Based on 10 cases diagnosed between 1997 and 2011, the mean annual incidence rate of TA was estimated at 0.7/million inhabitants (95% CI 0.3–1.2) and 1.5/million among women (95% CI 0.6–2.4). The mean annual incidence rate among population aged ≤ 40 years (based on 8 cases) was 1.1/million inhabitants (95% CI 0.3–1.9) and 2.3/million among women (95% CI 0.7–3.9). The corresponding incidence rate among patients of Swedish ancestry (based on 7 incident cases) was 0.6/million (95% CI 0.2–1.1) and among patients of non-Swedish ancestry was 1.4/million (95% CI 0–2.9).

*Point prevalence*. As of January 1, 2012, all 13 patients were alive and living within the study area. The point prevalence was estimated to 13.2/million inhabitants (95% CI 6.0–20.4). The corresponding prevalence among women was 26.2/million (95% CI 11.9–40.4). The corresponding point prevalence among patients of Swedish ancestry (based on 8 prevalent cases) was 10.2/million (95% CI 3.1–17.2) and among patients of non-Swedish ancestry was 25.5/million (95% CI 3.2–47.9).

*Table 1*. Clinical and laboratory characteristics of 13 patients with Takayasu arteritis from time of diagnosis. Values are median (IQR) or n (%) unless otherwise specified.

| Features/laboratory data | Value |
|---|---|
| Age at diagnosis, yrs | 23 (16–38) |
| Female:male | 13:0 |
| Diagnosis delay, mos | 12 (5–18) |
| Race | |
| Scandinavian | 9 (69) |
| Arab | 2 (15) |
| Asian | 1 (8) |
| African | 1 (8) |
| No. ACR criteria | 4 (3–6) |
| Main clinical features at diagnosis | |
| Constitutional symptoms | |
| Arthralgia/myalgia | 4 (31) |
| Fever | 3 (23) |
| Headache/fatigue | 7 (54) |
| Claudication | 6 (46) |
| Nonspecific chest or abdominal pain | 4 (31) |
| Absent peripheral pulses/murmur | 5 (38) |
| Hypertension | 5 (38) |
| Preeclampsia | 1 (8) |
| Episceleritis | 1 (8) |
| Main laboratory features at diagnosis | |
| Hemoglobin, g/dl | 11.6 (10.6–12.0) |
| White blood cells, × $10^9$/l | 9 (7–10) |
| Platelets count, × $10^9$/l | 420 (320–598) |
| C-reactive protein, mg/dl | 5.65 (1.6–9.57) |
| Erythrocyte sedimentation rate, mm/h | 55 (34–76.2) |
| Creatinine, mg/dl | 0.70 (0.51–0.83) |

ACR: American College of Rheumatology; IQR: interquartile range.

*The distribution of arterial lesions*. Data on radiological investigations were available for all patients. Vessel abnormalities included significant stenosis, occlusion, dilatations, aneurysm, and signs of blood vessel inflammation detected by positron emission tomography-computed tomography (PET-CT). The most common arterial territories affected were the left subclavian, descending aorta, left carotid, and right subclavian arteries (Table 2). Other affected arterial territories included the iliac and brachiocephalic arteries (2 patients each), and the inferior mesenteric, pulmonary, coronary, common carotid arteries, and ascending aorta (1 patient each).

*The treatment at diagnosis*. Seven patients were treated with a combination of corticosteroids and disease-modifying antirheumatic drugs (DMARD): 3 methotrexate (MTX; 1 patient in combination with etanercept), 3 cyclophosphamide, and 1 azathioprine. Three patients received corticosteroids as the only therapy. One patient was treated with an antihypertensive drug and surgical intervention (bilateral renal artery bypass surgery). For 2 patients, no data on treatment were available.

*The treatment at last followup*. Five patients received biologics (2 rituximab, 3 infliximab) in combination with DMARD (4 MTX, 1 azathioprine). Two patients were treated with biologics (1 infliximab, 1 tocilizumab) in combination with prednisolone. One patient was treated with MTX only, 3 patients were receiving low-dose prednisolone, and the remaining 2 patients were not taking any treatments. Indications for adding biologics were either disease progression or persistent disease activity despite treatment with corticosteroids and/or traditional DMARD.

*The outcome*. All patients were followed from time of diagnosis to December 1, 2013. The median time of followup was 9 years (IQR 4–17.5). No deaths occurred during the followup time.

*Organ damage*. The median VDI total score at last assessment was 5 for all patients (IQR 3.5–7). Eight patients (62%) had severe organ damage and 2 developed critical damage. Treatment-related damage was recorded in 3 patients (23%; Table 3). The most common organ systems affected by damage were the peripheral vascular (n = 13, 100%), followed by the cardiovascular (n = 10, 77%). Eight patients were affected by other damage/drug reactions (62%) and 3 were affected by gastrointestinal damage (23%). All items of damage registered in all patients are listed in Table 3.

*Surgical vascular interventions*. Five patients (38%) had a total of 8 surgical and 2 endovascular interventions during the followup time. The following vascular surgeries had been performed: carotid-subclavian bypass surgery (n = 1); bilateral renal artery bypass surgery (n = 1); subclavian artery graft (n = 2); carotid artery aneurysm stent operation (n = 2); and the reconstructive major surgery of ascending aorta with reimplantation of major vessels including the left subclavian artery, carotid communis, and the brachiocephalic artery (n = 1). The following endovascular procedures had been performed: carotid surgery and percutaneous transluminal angioplasty of the renal artery (n = 1), and of the subclavian artery (n = 1).

*Pregnancies*. A total of 23 pregnancies in 10 women were registered from case records. Sixteen pregnancies occurred before the TA diagnosis had been made and resulted in 15 live births (94%) and 1 abortion. Seven pregnancies occurred after the diagnosis of TA and resulted in 5 live births (71%); 3 patients underwent cesarean delivery, and 2 had normal vaginal delivery. One of the pregnancies after the diagnosis of TA was complicated by preeclampsia that was treated successfully, and a live cesarean birth followed. The other 2 pregnancies ended with 2 abortions, 1 of which was at the request of the patient. Among the 10 patients with pregnancies, 5 had lesions affecting vessels above and under the diaphragm (renal artery in 4 patients, superior mesenteric artery in 2, splenic artery in 2, inferior mesenteric artery in 1, celiac artery in 1, and iliac artery in 1).

*Table 2*. Distribution of arterial involvement in 13 patients with TA from southern Sweden compared with selected studies from different regions of the world. Values are %.

| Artery | This Study | Italy[15] | Greece[16] | Turkey[17] | Iran[18] | US[19] |
|---|---|---|---|---|---|---|
| Subclavian artery | 85 | | 97 | 76 | | |
| Left subclavian artery | 77 | 66 | 93 | | 47 | 69 |
| Right subclavian artery | 54 | 52 | 62 | | 27 | 40 |
| Descending aorta* | 70 | 39 | 72 | 22 | 40 | 37 |
| Carotid artery | 62 | | 91 | 52 | | |
| Left carotid artery | 54 | 44 | 82 | | 27 | 37 |
| Right carotid artery | 39 | 36 | 68 | | 20 | 25 |
| Renal artery** | 46 | | 63 | 26 | 13 | |
| Left renal artery | 8 | 34 | 57 | | | 15 |
| Right renal artery | 8 | 30 | 42 | | | 16 |
| Vertebral artery | 54 | | 48 | 19 | | |
| Left vertebral artery | 39 | 13 | 35 | | | 13 |
| Right vertebral artery | 39 | 12 | 22 | | | 12 |
| Superior mesenteric artery | 31 | 32 | 19 | 12 | | |
| Splenic artery | 31 | | | | | |
| Axillary artery | 23 | | | 11 | | |
| Coeliac artery | 23 | 21 | 33 | | | |

* Reported as abdominal aorta. ** Bilateral renal artery involvements in 4 patients in this study (31%). TA: Takayasu arteritis.

*Table 3*. Permanent organ damage according to the VDI in 13 patients with TA. Values are n (%). VDI items not registered in our patients are not shown in this table.

| Items of Damage | No. Patients |
|---|---|
| Major vessel stenosis[1] | 13 (100) |
| Claudication[1] | 8 (62) |
| Absent peripheral pulses | 7 (54) |
| Valvular disease | 6 (46) |
| Hypertension | 6 (46) |
| Mesenteric insufficiency/pancreatitis | 3 (23) |
| Angina/CABG[1] | 2 (15) |
| Myocardial infarction[1] | 2 (15) |
| Skin ulcers | 1 (8) |
| Avascular necrosis[2] | 1 (8) |
| Cataract[2] | 1 (8) |
| Impaired pulmonary test[3] | 1 (8) |
| Gut infarction/resection[3] | 1 (8) |
| Diabetes[2] | 1 (8) |
| Other damage/drug reaction | 8 (62) |

[1] Major vascular damage. [2] Treatment-related damage. [3] Critical damage. VDI: vasculitis damage index; TA: Takayasu arteritis; CABG: coronary artery bypass graft.

## DISCUSSION

To our knowledge, this is the first population-based report on epidemiology and clinical presentation of TA in Sweden. The previous data on the prevalence of TA, 6.4/million, originated from a tertiary referral center in Uppsala, Sweden, in 1983, and were based on the hospital discharge registry of 15 patients from the referral area[9]. The prevalence in our study is 2-fold higher than in previously reported Swedish data, though the comparison is difficult because of differences in the methodological approach in these studies. We present the first annual incidence estimates from Sweden, 0.7 per million, that are comparable with data from Denmark and the United Kingdom[3,4]. The study areas in our study and the Danish one are comparable in demographic and geographic characteristics, indicating that the incidence data presented in these 2 Scandinavian studies are reliable. There is only a limited amount of data on the prevalence of TA around the world[20], with the highest reported prevalence of 40/million in Japan[8]. Our prevalence figure of 13.4 per million is the highest reported in Europe. Cases in our study were retrieved by review of the ICD-10 codes for TA and aortitis in a large number of departments and all private rheumatology clinics in the area. We therefore believe that an explanation for our high prevalence figure is the well-established case retrieval strategy, as described[21]. In addition, a higher rate of immigration and excellent survival may partially explain our prevalence rate. However, no statistically significant differences were found when comparing the incidence and prevalence of TA among the population of Swedish ancestry to those born outside Sweden.

TA predominantly affects women. In our study, as well as in the previous Swedish report[9], all patients were women. In other European and North American studies, the proportion of female patients was between 84% and 93%[3,4,15,17], and was even lower in Kuwait (62%)[6] and in Iran (73%)[18]. It is not known whether these variations in sex distribution depend on methodological issues because some reports originated from epidemiological studies while others came from tertiary centers.

The median time of diagnosis delay in our study was 12 months, which is comparable to 14 months in Iran[18] and 15.5 months in Italy[15], but shorter than the 24 months in Greece[16]. The diagnosis delay is difficult to define because it is not

known when the disease starts. Still, for a considerable fraction of patients, TA presents with the vascular damage manifested by claudication and the absence of peripheral pulses. The lack of specific clinical characteristics early in the disease course, the rarity of the disease, and hence the low degree of awareness among physicians may contribute to the long diagnosis delay for TA[16,22]. The increasing use of PET-CT scan nowadays in the diagnostic workup will probably result in an earlier diagnosis in the future for patients with TA. An international multicenter effort is ongoing to establish diagnostic and classification criteria for primary vasculitis syndromes, including TA, integrating newer radiologic modalities such as PET-CT in the diagnosis of TA[23].

The most common vascular territory affected during the disease course was the left subclavian artery, similar to findings in other studies (Table 2). There are variations in reported frequencies of involvement of other vessels. However, these differences may be because of variations in the number of examinations available to each study or because data are extracted from different cohorts of patients.

The survival in our patients was excellent with no mortality during a median followup of 9 years. This finding is in line with other European studies[3,16]. There are no available outcome measures developed specifically for TA, although there are ongoing efforts and plans to address this important issue[24]. In our study, we applied the VDI to our cohort. All patients in our study experienced irreversible organ damage at least once and 62% had severe organ damage as defined by the VDI[14]. Comparison of the damage data we presented in our study is not possible because there are no published data specifically on the VDI in TA[25].

In our study, 38% of patients underwent endovascular or surgical vascular interventions because of TA during followup, comparable to what has been reported previously[19,26,27].

We present data on pregnancy and its outcome. In general, the fertility rate of women with TA in our study is similar to that of the average Swedish woman[12]. The outcome of pregnancies was favorable compared with other published data on pregnancy in TA[28,29]. One patient developed severe hypertension and preeclampsia that was successfully managed, and she delivered a healthy child. None of our patients developed severe valvular disease or heart failure during the course of pregnancy. Although it is difficult to speculate on the reason for these complications, in this study the rate of successful pregnancies decreased from 94% before the diagnosis of TA to 73% among the pregnancies that took place after the diagnosis. The effects of vascular disease, hypertension, and treatment used in TA could have contributed to this complication rate.

The limitation of our study is its retrospective nature, sometimes resulting in incomplete data. However, the strengths are that our study is population-based, presenting both an epidemiologic update as well as a description of clinical characteristics, vascular involvement, and prognosis in terms of comorbidities as presented by organ damage. To the best of our knowledge, this is the first presentation of pregnancy outcome in TA in a North European population.

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